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Case Report

Congenital pulmonary hernia secondary to absence of ribs

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ABSTRACT

Congenital lung hernia is extremely rare with less than 50 reported cases.

We report two cases of lung hernia, secondary to congenital absence of ribs – A 4-year-old girl without any antecedent history of chronic cough or chest trauma presenting with a left lower lobe hernia secondary to an absent left 9th rib; a 7 month-old girl with recurrent pneumonia presenting with severe respiratory distress, fever and severe malnourishment, found to have absent 6th-9th ribs on right side with associated liver and lung herniation. The older girl has been kept on observation without surgery but the infant expired within 48 hours of admission due to respiratory failure.

The clinical scenario is a rarity and can be managed conservatively in most cases. Surgical treatment should be considered in symptomatic patients and in those with severe complications. Repair for cosmetic reasons is sometimes justified.

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1. Introduction

Hernia is a general term used to describe an abnormal protrusion of an organ through the structure or muscle that usually contains it.

Pulmonary or lung hernia (also known as a pneumocele) is an extremely rare entity. It refers to part of a lung bulging through a weak spot in the chest wall. In most of the reported patients, lung hernias are the result of injury or trauma to the chest, such as a fall, road traffic injury or surgery. Congenital lung hernia is extremely rare with less than 50 cases reported till date.

We report two such cases of lung hernia, secondary to congenital absence of ribs, along with a review of the literature.

2. Case Reports

2.1. Case 1

A 4-year-old girl with no history of chronic cough or any chest trauma came with a swelling on the left lower chest. She had no associated symptoms like pain, cough, respiratory distress, etc.

2.1.1. Past history was unremarkable.

On examination, it was a 4x4 cm globular lump in the left lower antero-lateral chest wall approximately 8-9 cm lower and lateral to the left nipple. (Figure 1) The swelling was soft, cystic with a smooth surface, reducible and moved in and out with respiration. The patient underwent a chest CT scan and a lung hernia was revealed along with an absent left 9th rib. Associated findings included a flattened 8th rib and bifid 10th rib on the left side. The ribs on the right side were all normal. No abnormalities were noted in the lung fields. (Figure 2)

The patient has been under observation and 6-monthly OPD follow-up for the last 16 months and is doing fine till date.

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Fig. 1: Clinical picture (Case 1)

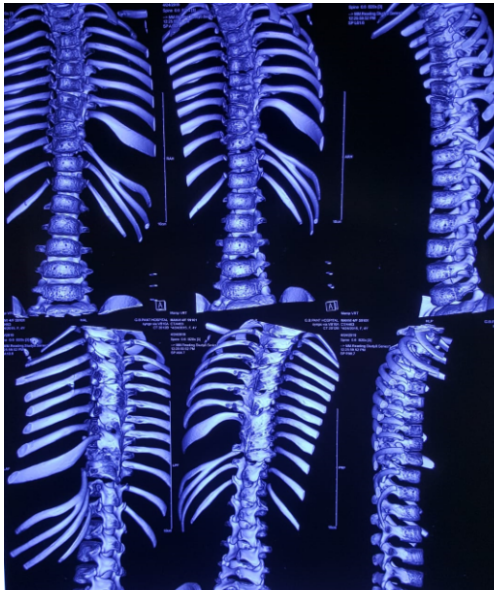


Fig. 2: CT scan (Case 1) showing rib absence and associated anomalies

2.2. Case 2

A 7 month-old girl with recurrent episodes of pneumonia since birth was brought with severe respiratory distress along with fever. She had 3-4 such similar episodes since birth and was severely malnourished.

On examination, the chest was asymmetrical with depression of the right antero-lateral chest wall below the nipple. (Figure 3) There was a bulge in the right lower chest with paradoxical movement on respiration. The postero-anterior Roentgenogram showed absence of the 6th-9th ribs and a bifid 10th rib. There was associated liver herniation. Lung herniation could not be appreciated on the Xray. The ribs on the left side were all normal. (Figure 4)

A CT scan was planned but could not be done as the patient had a downhill course after admission and expired within 48 hours.



Fig. 3: Clinical picture (Case 2)



Fig. 4: Chest X-ray (Case 2) showing absence of ribs and soft tissue herniation

3. Discussion

The occurrence of hernial protrusions of the lung is exceedingly rare and the literature on this topic is scarce. It was first described by Roland in 1499.¹

The usual presentation is in the supraclavicular, parasternal or paravertebral region or in an area of the chest wall with a congenital absence of rib(s). Any situation causing an increased intrathoracic pressure may result in a lung hernia if there is an associated anatomic weakness in the thorax.²

The most widely accepted classification is that of Morel-Lavalle³ based on the anatomic location (cervical, thoracic, or diaphragmatic). These groups are stratified further according to the presumed etiology as congenital or acquired; the latter group contains the traumatic, “spontaneous,” pathological and the post-surgical varieties. (Table 1)

Table 1: Classification of lung hernias

Classification of lung hernias	
Anatomic location	<ul style="list-style-type: none"> • Cervical • Thoracic • Diaphragmatic • Mediastinal
Etiology	<ul style="list-style-type: none"> • Congenital • Acquired Traumatic Spontaneous Pathological Post-surgical

Patients at the highest risk of a lung hernia are adults with elevated intra-thoracic pressure, such as patients with morbid obesity or end-stage chronic obstructive pulmonary disease.⁴ Other risk factors include tissue weakness or poor healing from malnutrition, steroids, diabetes, or other comorbidities. Lung herniations encountered in the pediatric age group are extremely rare and are mostly congenital in origin; acquired or traumatic hernias are rare.

Congenital hernias result from the attenuation of the endothoracic fascia. They occur either at the thoracic inlet or at the intercostal spaces, where weakness of the fascia is usually combined with absence of the intercostal muscles.⁵ Approximately 18-24% of reported cases of lung hernia are congenital.^{2,6} Approximately 60% of congenital lung herniations are cervical, the remaining 40% being parasternal or paravertebral herniations or herniations through the chest wall at the site of a congenital absence of ribs. Herniation of lung through the diaphragm is most unlikely because intraabdominal pressure is usually higher than intrathoracic pressure. It has only been reported on one occasion by Beale in 1882.⁷

The location of a lung hernia is usually anterior, especially in the lower intercostal spaces; the next most common position seems to be lateral, with the rarest position being posterior. This is due to anatomic conditions; in fact, the anterior inferior part of the chest has much less muscular support than the anterior superior part, the sides, or the posterior part, and the lower costal cartilages are more widely separated than the rest.⁸

Pulmonary hernia can be asymptomatic. Deformities of the inferior ribs usually form a ‘lung hernia’ that produces an unstable chest, leading to recurrent respiratory infections and paradoxical respiratory movements.⁹ Standard posterior-anterior X-ray of the chest usually fails

to demonstrate a lung hernia, but do reveal the site of rib deficiencies if they exist. Oblique or lateral projections with the involved area in profile are diagnostic. Forced expiration with straining against a closed glottis will cause protrusion of the lung into the hernial sac and is an important manoeuvre to identify this lesion. The diagnosis should be confirmed by a computed tomography (CT scan) of the thorax to assess the exact location and size of the defect.¹⁰

Treatment may be conservative or surgical, but as experience is limited, no long-term results have been reported. Most children with lung herniations require nothing beyond reassurance of the parents, emphasizing the lack of complications or dangers associated with these defects. However, with a progressive increase in size, a history of incarceration, unrelenting respiratory distress or if a severe cosmetic problem is present, consideration must be given to surgical repair of these lesions. They rarely heal spontaneously (Felliers¹¹ reported a spontaneous cure of bilateral cervical lung hernia in a child, the only spontaneous cure reported in literature). One patient of lung hernia reported by Leary in 1966¹² died of bronchopneumonia at the age of 2 months similar to the case we presented. Our other patient is under observation for the last 16 months and has not developed any symptoms till date.

In conclusion, the clinical scenario is a rarity and can be managed conservatively in most cases. Surgical treatment should be considered in symptomatic patients and in those with severe complications. Repair for cosmetic reasons is sometimes justified.

4. Disclosure Statement

Appropriate written informed consent was obtained from the parents for publication of this case report and accompanying images.

5. Conflict of Interest

The authors declare that there are no conflicts of interest in this paper.

6. Source of Funding

None.

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